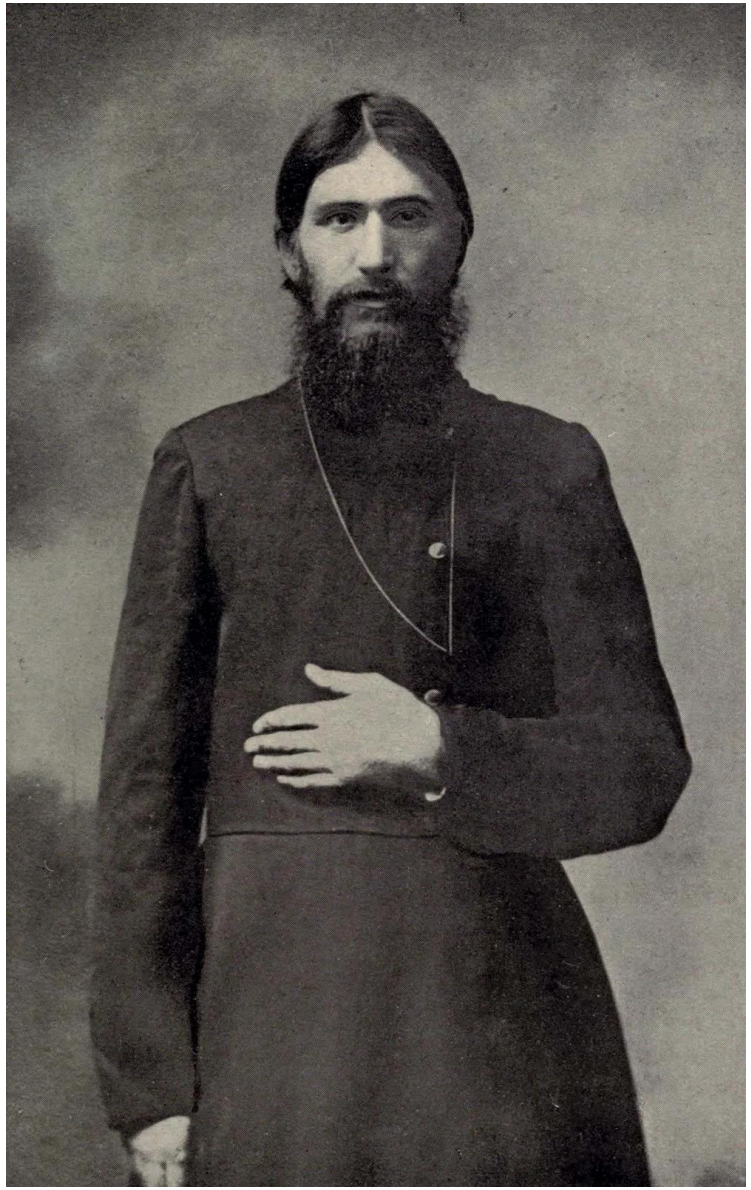
Haemophilia: Pregnancy and Birth

Dr Iniyaval Thevathasan

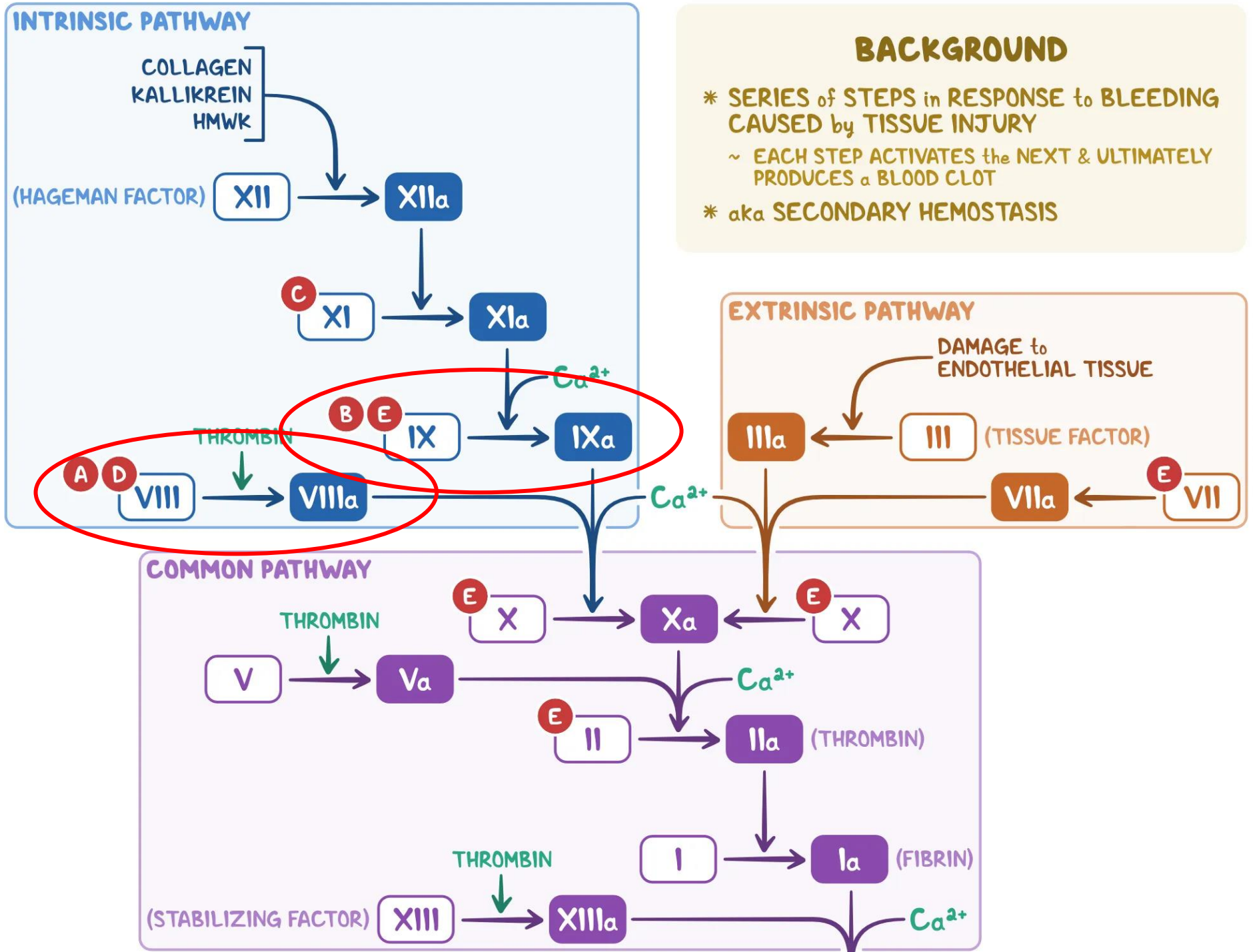
Obstetrician and Maternal Fetal Medicine Subspecialist

MBBS(Hons), FRANZCOG, CMFM, DDU(O&G)


E.N.A.
BOISSANNAS
ET EGGLER



But what if, Alexandra had access to modern obstetric care?


INTRINSIC PATHWAY
COLLAGEN
KALLIKREIN
HMWK
(HAGEMAN FACTOR)
XII
XIIa
C
XI
XIa
Ca2+
B
E
IX
IXa
THROMBIN
A
D
VIII
VIIIa
BACKGROUND
* SERIES of STEPS in RESPONSE to BLEEDING CAUSED by TISSUE INJURY
~ EACH STEP ACTIVATES the NEXT & ULTIMATELY PRODUCES a BLOOD CLOT
* aka SECONDARY HEMOSTASIS
EXTRINSIC PATHWAY
DAMAGE to ENDOTHELIAL TISSUE
IIIa
III
(TISSUE FACTOR)
E
VIIa
E
VII
Ca2+
COMMON PATHWAY
THROMBIN
E
X
Xa
E
X
V
Va
Ca2+
E
II
IIa
(THROMBIN)
THROMBIN
I
Ia
(FIBRIN)
(STABILIZING FACTOR)
XIII
XIIIa
Ca2+

Physiology

- Normal Factor VIII and IX levels
  - 50-200 IU/dL
- Physiological response of coagulation factors to pregnancy
  - Factor VIII increases
  - No change to Factor IX
- Severity
  - Mild – 5-40 IU/dL
  - Moderate – 1-5 IU/dL
  - Severe - <1 IU/dL
- Symptomatic carriership
  - < 40 IU/dL with bleeding phenotype
- Asymptomatic carrier
  - <40 IU/dL without bleeding phenotype

Dunkley S et al. Med J Aust 2019.

Why is haemophilia important to obstetricians?

Safer Care Victoria 2019
D'Orion R Haemophilia 2021

Pre-pregnancy counselling

- Multidisciplinary counselling
- Assess factor levels
- Historical response to DDAVP
- Fertility options
- Set expectations

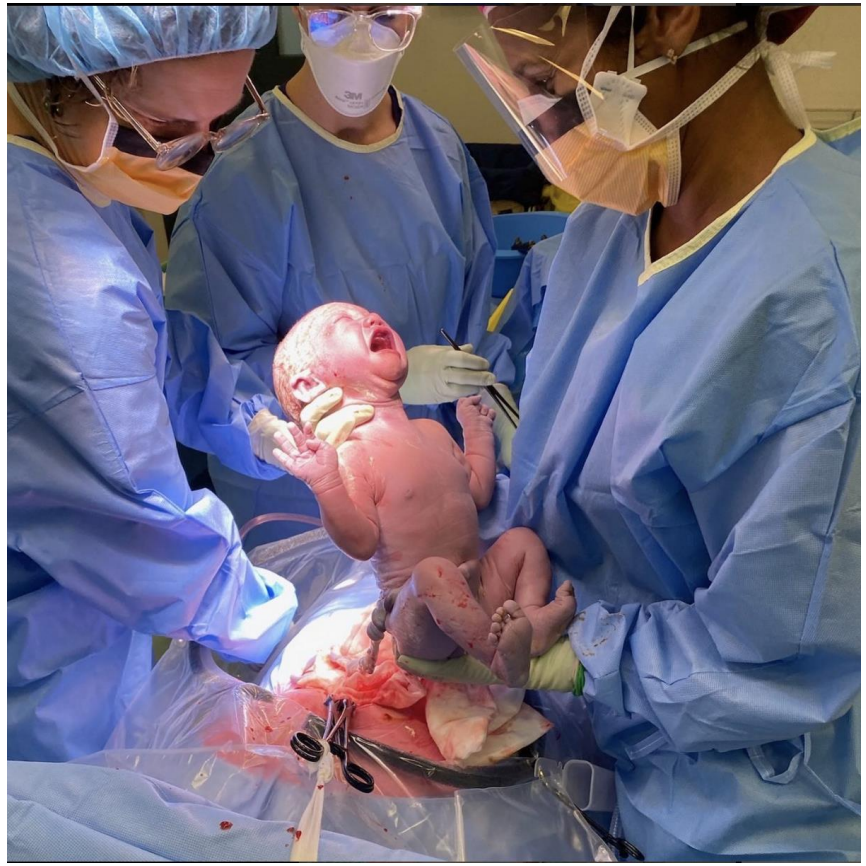
Routine pre-pregnancy counselling

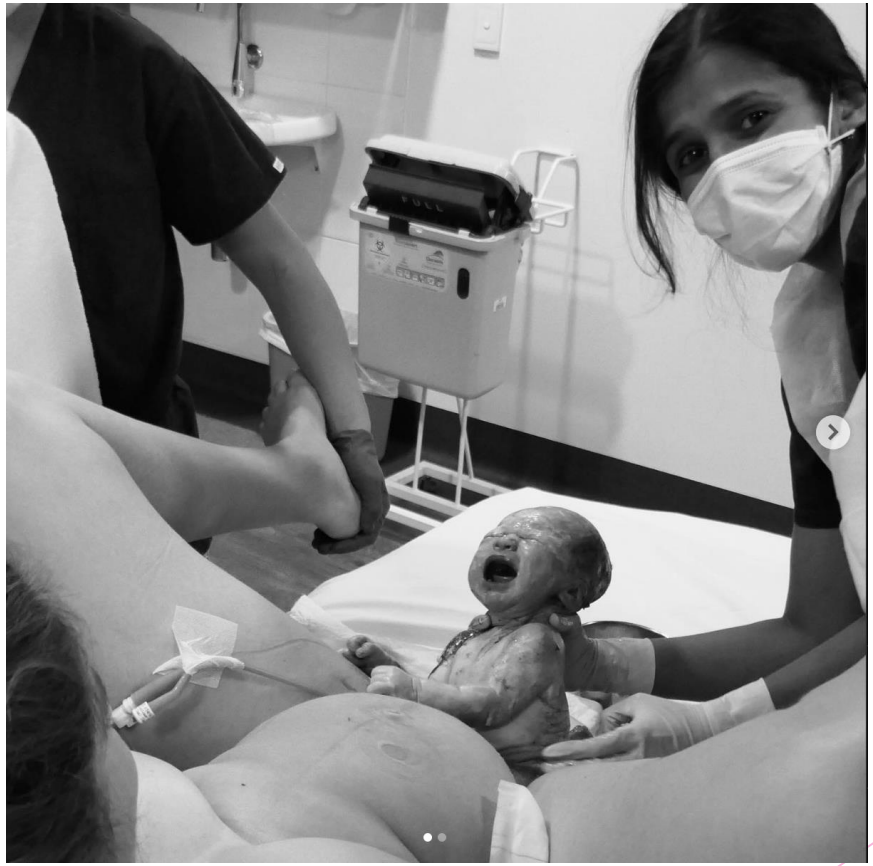
- Folic acid
- Recessive carrier screening
- Immunity to vaccine preventable diseases
- Medication optimization and assessment of other comorbidities
- Routine screening tests
- Diet, exercise and sound mental health

Antenatal Care

- Multidisciplinary visits
- Consideration of antenatal testing
- Measurement of factor levels
- Review PPH risk factors
- Anaesthetic review
- Paediatric review

Mode of Delivery





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The argument for caesarean section

- Maternal factors
  - Sense of control
  - Choice
  - PPH rates not increased
- Fetal factors
  - Increased risk of intracranial haemorrhage with vaginal delivery
  - Operative vaginal delivery may be unavoidable

Davies J et al Haemophilia 2016
Kulkarrni R et al Haemophilia 2009

What we all agree on

- Operative vaginal delivery should be avoided
  - If unavoidable, forceps over vacuum
  - Experienced obstetrician
- In women carrying a female fetus, mode of delivery on obstetric indications
  - Remains a risk of non severe bleeding
- A priori risk and shared decision making should be factors in determining mode of delivery
- Don’t be an obstetric cowgirl(boy)

Dunkley S et al. Med J Aust 2019.
Pacheo L et al. Obst Gynaecol 2023.

Intrapartum care

- Large bore IV cannulas, X-Match
- Factor replacement ± TXA ± DDAVP
- Avoidable risks for PPH
- Active management of the third stage

Immediate postpartum care

- Maintain normal range factor levels
- Regional anaesthesia considerations
- Postpartum TXA
- Avoid LMWH and NSAIDs
- Neonatal paediatric review

Longer term postpartum care

- Assess and treat postpartum bleeding
- Breastfeeding
- Mental health
- Contraception
- Paediatric follow up
- Plan for the next pregnancy

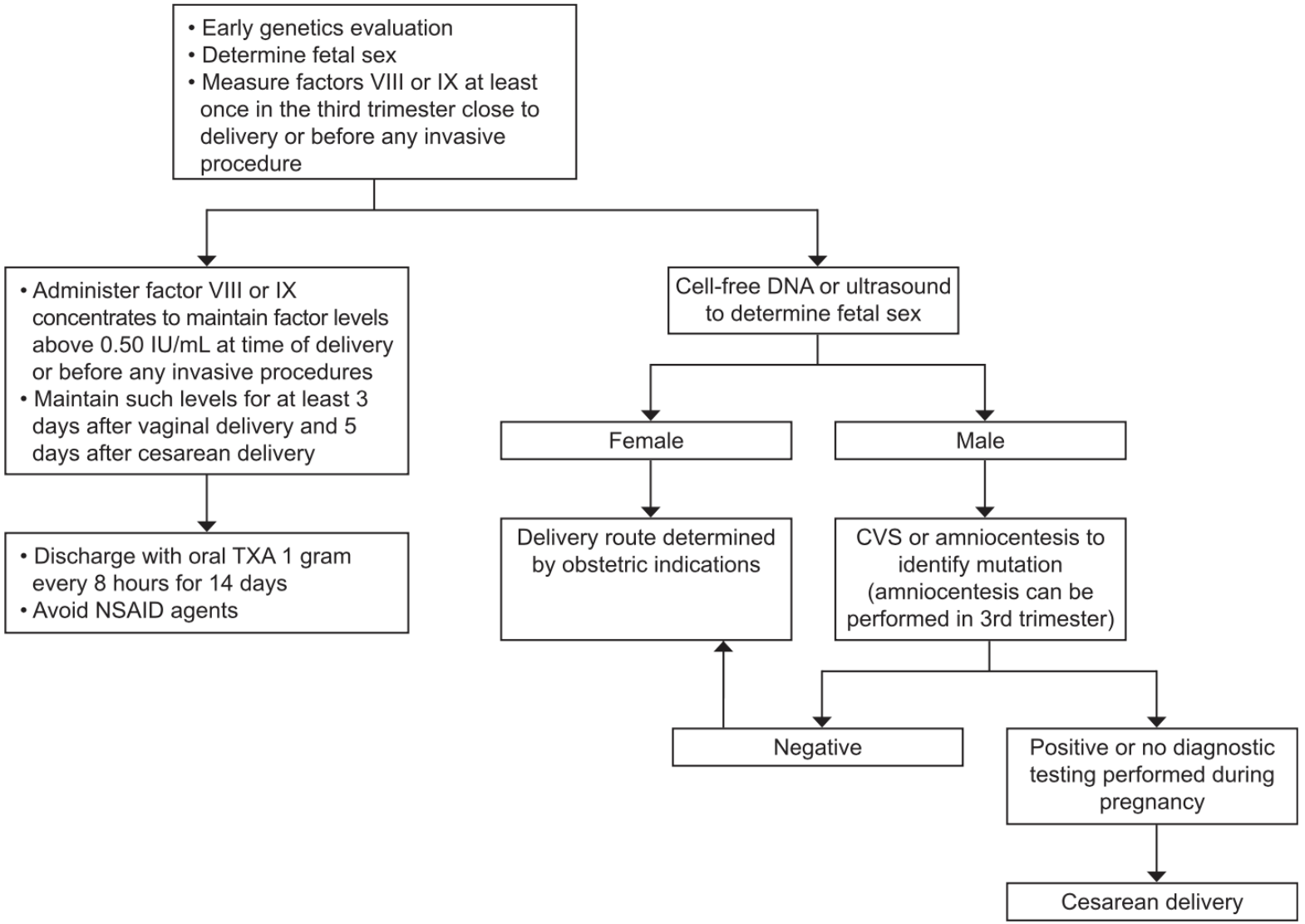

Fig. 3. Management of hemophilia during pregnancy. TXA, tranexamic acid; NSAID, nonsteroidal anti-inflammatory drug; CVS, chorionic villus sampling.

Pacheco. Inherited Bleeding Disorders in Pregnancy. Obstet Gynecol 2023.

Key Points

- Women with haemophilia can have successful pregnancies and healthy families
- Pre-pregnancy counselling offers women choice and optimizes pregnancy outcomes
- Multidisciplinary care and careful planning is key
- Mode of delivery remains controversial but operative vaginal delivery should be avoided
- Don’t forget the routine care!

Questions?

No questions about hypothetical historical accuracy please.

Dr Iniyaval Thevathasan

Melbourne MFM, (03) 9411 7474

admin@melbournemfm.com.au

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